LETTERS TO THE EDITOR

‘Lupus lymphadenitis’ simulating a strangulated femoral hernia in a patient with mixed connective tissue disease

SIR: Lymphadenopathy is a common finding in both systemic lupus erythematosus (SLE) and mixed connective tissue disease (MCTD) and is often associated with disease activity. Aneurysm formation may be a feature of the degree of lymphadenopathy sufficient to cause concern over the possibility of primary lymphatic disease, an acute presentation with localized lymphadenopathy in the absence of other systemic features is exceptional. We report a patient in whom the clinical presentation suggested the diagnosis of a strangulated femoral hernia, but in whom the true diagnosis, on histological examination, was lupus lymphadenitis.

A 25 year old woman presented with a five day history of a painful lump in her right groin associated with a raised temperature and general malaise. She had had some intermittent swelling in her right groin five months earlier. In 1983 she had had a partial thyroidectomy for thyrototoxicosis and in 1986 she was diagnosed as having SLE when she presented with general malaise, weight loss, Raynaud’s phenomenon, and a cough. She was found to have diffuse hypergammaglobulinaemia, a negative rheumatoid factor, a positive antinuclear factor (titre 1/80, speckled pattern), perinuclear anti-DNA binding (titre 1/5000), and a positive extractable nuclear antigen (titre 1/732). She then had two episodes of myopericarditis, both of which responded well to intravenous methylprednisolone. She has now developed scleroderma changes affecting her hands and face and oesophageal dysmotility. She has never had any clinical features to suggest Sjögren’s syndrome. Her autoantibody pattern has changed with the development of antihistone antibodies and the loss of DNA binding, suggesting progression to MCTD with features of scleroderma. Throughout this time her maintenance dose of oral prednisolone and azathioprine has been unchanged. On examination she was cushingoid with facial telangiectasia, microstomia, and sclerodermatous changes in her hands. She was afebrile, but had a tender mass in her right groin measuring 3×3 cm, which was thought to be a strangulated femoral hernia. At operation she was found to have several enlarged lymph nodes below the right inguinal ligament. She made a good postoperative recovery and has since remained well with her usual maintenance dose of prednisolone 7 mg/day and azathioprine 50 mg/day. Histology of the lymph nodes was characteristic of lupus lymphadenitis showing necrosis (figure) with nuclear debris underlying phagocytes in one lymph node and both centroblastic and immunoblastic responses in adjacent nodes. Lymphoid follicles and generalised lymphadenopathy occurs in about half of patients with SLE at some stage during their disease1 and in about a third of patients with MCTD.3 In SLE superficial nodes, particularly those in the neck, are commonly affected. Necropsy studies, however, commonly show enlargement of mesenteric, tracheobronchial, and retroperitoneal nodes, suggesting more generalised involvement. The pathology of lymph nodes in SLE is well reported and characteristic,4 though the pathogenesis remains unknown.4 Afebrile lymph node showing marked necrosis. (Haematoxylin and eosin.)

Section of lymph node showing marked necrosis. (Haematoxylin and eosin.)

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Raised concentrations of von Willebrand factor antigen in systemic sclerosis

SIR: von Willebrand factor antigen is the antigenic component of von Willebrand factor, and circulating concentrations are raised in vasculitis and systemic sclerosis, reflecting damage to endothelial cells. In systemic sclerosis concentrations of von Willebrand factor antigen correlate with a risk of mortality1 and with the extent of visceral disease—that is, involvement of heart, lung, muscle, etc.2 Neither of these reports directly considered the question of concentrations of von Willebrand factor antigen in the two major subgroups of systemic sclerosis—the limited cutaneous variant (CREST) and the diffuse variant.

We therefore studied 14 patients with clearly defined CREST but no proximal skin disease and six with the clearly defined diffuse variant with proximal scleroderma,3 and compared them with 20 apparently healthy laboratory and hospital staff as controls matched for age and sex. von Willebrand factor antigen in plasma was measured by a standard enzyme linked immunosorbent assay (ELISA).4 Results are shown in the figure. von Willebrand factor antigen was raised in patients with systemic sclerosis relative to normal controls (median 1500 IU/I, controls 800 IU/I, p<0.01, Mann-Whitney U test used throughout). Analysis of variance showed that both the diffuse variant (1770 IU/I, p<0.01) and the CREST variant (1150 IU/I, p<0.05) had raised concentrations of von Willebrand factor antigen, which were higher in the patients with the diffuse variant than in those with the CREST variant (p<0.01).

These data confirm and extend previous reports about von Willebrand factor antigen in systemic sclerosis. Sharman and colleagues studied 34 patients with systemic sclerosis, of whom seven died (mean von Willebrand factor antigen 2980 IU/I, in the surviving group 1450 IU/I).5 Creaves et al found a mean von Willebrand factor antigen of 1730 IU/I in 16 patients with severe disease and 1030 IU/I in 12 patients with mild disease.6 Our finding